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THE BRITISH JOURNAL
OF
CHILDREN'S
DISEASES.
EDITED BY GEORGE CARPENTER, M.D.

REPRINT

PRINTED AND PUBLISHED BY ADLARD & SON,
BARTHOLOMEW CLOSE, LONDON, E.C.

ON SOME OF THE DISEASES AND INJURIES OF THE EYE PECULIAR TO CHILDREN.

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Reprinted from 'The British Journal of Children's Diseases,' 1905.

ON SOME OF THE DISEASES AND INJURIES OF THE EYE PECULIAR TO CHILDREN.*

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Many of the abnormal conditions of the eye in childhood result from developmental anomalies, but these for the most part I shall refrain from touching upon, since they will be described during the present course by Mr. E. Treacher Collins. My remarks will be limited to certain diseases and injuries of the eye met with exclusively, or almost exclusively, in children. You will therefore perceive that my subject is necessarily a somewhat small one. At the same time, as I hope to show, it is one of considerable practical interest and of no little importance.

A few cursory remarks with regard to the examination of children's eyes may serve as an introduction to the actual description of disease.

To estimate the sight in an infant is by no means so simple a matter as might at first be thought. If the child is in hospital and has been there for some little time, valuable information may be obtained from the nurse as regards the recognition of food, of parents, and so forth. The observations of a moderately intelligent mother, too, are often helpful. They are likely to be correct in case the sight has failed after being good, but are often at fault, on the other hand, when the infant is believed never to have seen properly. The test usually applied by the surgeon is to ascertain whether an infant turns its eyes towards a mirror when light from the latter is thrown into the eyes in a darkened room. According to comparative tests conducted at Queen Charlotte's Hospital, London, even a baby but a few days old will respond to the "flash-test," but as the faculty of sight, and especially of fixation, is slowly acquired, the baby must not be expected to follow the movements of the light until he has attained an age of several weeks. It would be a perfectly legitimate generalisation to say that all babies of three months old and upwards who do not respond to this test are either blind or imbecile or actually idiotic.

* A lecture delivered on July the 11th, 1905, in the post-graduate course of ophthalmology in the University of Oxford.

Points to bear in mind in attempting to estimate the sight of little children are: (1) The presence or absence of rolling movements of the eyes, nystagmus, or squint. (2) The activity or otherwise of the pupils to light. (3) Any departure from normal as regards the size or conformation of the skull, a point in connection with which I may recall to your mind the fact that at birth the normal skull has a circumference of about 13 inches, at 1 year of 18 inches, and at 5 years of 20 inches. (4) The mental and bodily development of the child. With regard to this point you should be familiar with what Dr. Robert Hutchison ('Lectures on Diseases of Children,' 1904, p. 17) has aptly called "milestones" in the development of the normal child. The most important of these are as follows: the baby begins to cut his teeth at 6 months, and has all his milk teeth at the age of 3 years. The anterior fontanelle should be closed between the 18th and the 24th month. The baby should hold his head up at the age of 3 or 4 months, sit up from 9 to 12 months, walk at 12 to 18 months, and talk at 2 years. It is also important that you should notice the presence of the so-called "stigmata of degeneration," of which, perhaps, the commonest are marked epicanthus, vaulted palate, asymmetrical skull, supernumerary auricles, webbed fingers, post-anal dimple, and epispadias.

In older children, up to the age of, say, 5 years, a quantitative test of sight can easily be applied by ascertaining the distance at which familiar objects, as toys, can be recognised. As children can usually count before they know their letters, a rough test may be made on Snellen's types by getting the child to count the number of letters in each line, and by then entering the vision in the ordinary way, of course noting how it was elicited. An even better plan is that devised by Mr. Claud Worth ('Squint,' London, 1905, 2nd edition, p. 85), who employs five ivory balls, which vary in size from half an inch to one and a half inches. After the child has been accustomed to the little balls by being allowed to handle them, they are thrown on the floor to a distance of six or seven yards, turn by turn, beginning with the largest. The child is then told to go and to pick them up, and, as Mr. Worth says, "It is easy to tell by the way in which the child runs for the ball whether he really sees it before he starts, or is only going to look for it." I have found Mr. Worth's plan succeed with most children who can walk—that is, after two years of age.

After five years of age most children of the better class may be tested in the ordinary way at Snellen's distant types. With hospital patients, however, unless they appear to be unusually intelligent or

something material depends upon the result, I do not usually test them on Snellen's types until they have reached the age of 7 years or upwards. In this connection it is well to know what position the child occupies at school, since, as a rule, it is not worth while to test an "infant," while "boys" and "girls," especially if over the first standard in an elementary school, may readily be tested. Roughly, you will remember that the first standard contains children of 7 to 8 years, the second those of 8 to 9 years, the third those of 9 to 10 years, and so on in proportion. A comparison of the child's age with the school standard, therefore, will not only afford a rough indication of the child's intelligence, but will also enable you to gauge the value to be attached to the estimation of the relative visual acuity as determined on the distance types. A good deal of time and of tact are called for in order to reach a true estimate of a child's relative visual acuity. Above all things, a child resents any appearance of hurry on the part of the examiner. Again, if he be hectored in the least, he will almost certainly fail to do justice to himself, and in that way convey a false estimate of his sight to the surgeon. It is better not to harry the child over somewhat difficult letters as "Z," "X," and "Q," but to allow him to slur them over, or even to miss them altogether. If the contrary course be adopted, he will often come to a full stop at the difficult letter, and will obstinately refuse to go any farther, even although with a little coaxing he will readily read two or three smaller lines.

Despite the sweet innocence of childhood—more often sung by poets than seen in the out-patient department of a children's hospital—care should always be taken to prevent deception, particularly in the case of children who squint; for example, the child should never be allowed to cover up one eye while the sight of the other is being taken.

In using the ophthalmoscope in adult patients we proceed with our investigation in a definite and regular order—first, examining the optic disc, then, the yellow spot region, next, the circumpapillary area, and, lastly, the peripheral parts of the fundus. In a young or fractious child this orderly sequence can seldom be followed. The region that first presents itself (usually the yellow spot region) should be scrutinised, and then we must, as far as possible, explore the other parts of the fundus, but without adopting any formal order in the process. In the case of children lying ill in bed, the difficulties of ophthalmoscopic examination are multiplied tenfold. The patients are sometimes too ill to be moved, and are often so irritable as to render satisfactory examination a matter of the greatest difficulty. Under such circumstances we have often to content ourselves with a

passing glimpse of the optic disc or of any morbid changes that may be present. It is a fortunate thing that some of the most important ophthalmoscopic changes—tubercle of the choroid in particular—are seldom found elsewhere than in the central and most accessible portion of the fundus oculi.

If children lie in a state of "coma-vigil" or are actually unconscious, the examination can be made without let or hindrance, although we may be compelled to introduce a speculum between the lids, and to draw the eyeball into any desired position by means of fixation-forceps.

Both the direct and the indirect methods of ophthalmoscopic examination are useful in investigating the eyes of children in bed, and, in my opinion, one should not be employed to the exclusion of the other. For this kind of work the electric ophthalmoscope (of which several good patterns are now on the market) furnishes considerable help, since as the source of light and the mirror practically coincide, no co-ordination on the part of the surgeon is needed. The instrument can scarcely be praised too highly for facilitating the examination of bedridden children, although its limitations in other directions will at once strike those who endeavour to use it in ordinary out-patient work. Whatever form of ophthalmoscope we employ, it is seldom, if ever, worth while to endeavour to make an examination with undilated pupil. The pupil should always be dilated as fully as may be with some agent, as a 5 per cent solution of euphthalmine or a 10 per cent. solution of mydrine, which acts little or not at all upon the accommodation. Atropine (grs. 2 or 4 to the ounce) may, of course, be used instead of the newer agents, but that involves as its necessary consequence a paralysis of the accommodation, which may be troublesome unless the child is too young to complain or too ill to use his eyes. There is, moreover, a more cogent objection to atropine, namely, that its effects take several days to pass away, and during that period the reactions of the pupil—often a point in the diagnosis of certain nervous disorders—cannot be estimated properly.

Finally, if the patient be extremely troublesome, or important issues hang upon the results of examination, personally I never hesitate to administer ethyl chloride, and after having introduced a speculum, to make the ophthalmoscopic examination. The anæsthetic in question, the action of which seldom lasts for more than two or three minutes, gives ample time for the examination to be made.

The *technique* of using the ophthalmoscope in older children does not differ materially from that adopted in grown-up people, except that the child should be steadied by kneeling upon a chair and by

being made to grasp the top rail of the chair with his hands. The services of a nurse will save the surgeon an infinity of trouble by keeping the child's head still during the examination. In the absence of help of this kind, the same object may be attained by grasping the lobe of the patient's ear between the finger and thumb of the disengaged hand during the direct examination.

It is advisable to have some definite object to which the child may look during the examination of the right or left eye, as the case may be. I find a couple of small glow lamps, one red and the other green, fixed on the wall at the far end of the room, useful. Failing some such simple device, a toy that rattles or jingles when shaken is useful in attracting a child's attention.

The examination of the field of vision with the perimeter seldom offers any difficulty if undertaken in a child of five years or upwards. It is advisable that this fact should be recognised more widely than appears to be the case, inasmuch as the diagnostic value of the field of vision in children sometimes cannot be over-estimated. Much as many children possess a visual acuity above the normal, so the limits of the visual field for white are often found to exceed the usual line.

The estimation of intra-ocular tension in babies is likely to yield most fallacious results unless a general anæsthetic is first administered for the purpose of doing away with muscular movements.

After the foregoing casual remarks, I pass forward to discuss the subject in hand, namely, diseases and injuries of the eye in childhood. In doing this I shall first take up affections of the lacrymal apparatus in children, and then congenital ophthalmia neonatorum. After that, "keratomalacia" and the mysterious disease known as buphthalmos or "infantile glaucoma" will occupy our attention. Then "deformities of the skull and defects in sight" will be discussed, to be followed by "nystagmus" and "amaurosis" in children, some "family affections of the eye" and "congenital word-blindness." My remarks will conclude with a description of certain injuries to the eye liable to be sustained by the infant during his passage into the world. I shall pass over in silence cataract (congenital and infantile), squint, eczematous ophthalmia, interstitial keratitis, spring catarrh, and glioma and pseudo-glioma, because the time at my disposal is too limited to allow me to deal adequately with those important subjects.

I would venture to digress from the subject so far as to insist upon the importance of general disease as a factor in the causation of affections of the eye in children. In this connection it is only

necessary to point to the influence of digestive disorders, of gonorrhœa, of tuberculosis, and, above and beyond all, of syphilis. Those who would deal successfully with the ocular diseases of infancy should therefore have something more than a mere passing acquaintance with the general ailments to which children are liable, a point, indeed, too obvious to be laboured.

Affections of the Lacrymal Apparatus.

Dacryoadenitis.—Inflammation of the lacrymal gland, dacryoadenitis, is rare in children. I have myself met with it under two conditions—(1) as a complication of epidemic parotitis, and (2) as an idiopathic inflammation.

I may briefly quote an example of each condition :

(1) George P—, aged 7 years, developed, on June the 10th, 1901, an ordinary attack of mumps, characterised by a slight rise in temperature (101° F.), and swelling of the right parotid gland. On June the 15th the other parotid was affected, the tumefaction of the first having in the meanwhile subsided almost completely. Three days later (June the 18th) the parotids were no longer enlarged, but the upper lid of the right eye was red and œdematous, and a somewhat tender lump was found to occupy the lacrymal fossa. The affected eye watered a good deal. On June the 20th the symptoms had been transferred to the other eye. The patient had recovered by June the 24th—*i. e.*, a fortnight after the parotitis had commenced.

(2) Aubrey D—, aged 3½ years, was first seen on April the 3rd, 1905. The history was that four days before admission into the Evelina Hospital the left upper lid had become red, painful, and swollen, and that a thick discharge had been noticed to issue from the eye. There was no history of mumps or of exposure to the infection of that disorder. Upon admission, the left upper eyelid was markedly swollen and discoloured, while the lower lid showed slighter changes of the same kind. There was a profuse discharge of thick pus from the eye. At first sight the case resembled one of gonorrhœal ophthalmia. But examination under a general anæsthetic showed that a hard nodule was present in the position of the lacrymal gland, whence it extended for some distance into the outer part of the œdematous upper eyelid. The upper and outer part of the ocular conjunctiva was chemotic, and the chemosis overlapped the cornea, which was clear and bright. No proptosis. Parotid glands not enlarged. Temperature 99·2° F. A narrow blade pushed through the upper lid into the nodule gave exit to no pus. Under hot boric fomentations the condition receded spontaneously, and the child was discharged well after a stay in hospital of thirteen days. During that period the general condition of the child was good, and the highest point reached by the temperature was 99·6° F. The child was brought to the out-patient department on May the 2nd, when it would have been difficult to tell from inspection which had been the affected eye.

Dacryocystitis.—For reasons that will appear immediately, it is desirable to distinguish between affections of the tear passages as such occur in children under one year of age and in those over that age.

(*a*) A baby, usually less than 6 months' old, is brought with the

statement that the eyes have been noticed to discharge either from or shortly after birth. On examination of the infant, a little plug of muco-pus can be seen to lie at the inner canthus or to glue the cilia together. There is no swelling of the eyelids, so that the baby can open his eyes freely; the eyeball is not bloodshot; the cornea is clear. Although the palpebral conjunctiva may be somewhat hyperæmic, yet it has never the thick, red, folded, and villous appearance so characteristic of ophthalmia neonatorum. It is the exception for an obvious swelling to exist in the region of the lacrymal sac—*i. e.*, at the side of the root of the nose, behind the tendo oculi. More commonly there is a slight, ill-defined fulness of the region in question. When firm digital pressure is made over the internal palpebral ligament muco-pus may be seen to exude from one or other punctum, and a notable amount of discharge may in that way often be squeezed into the conjunctival sinus. A less frequent event is for compression to give rise to an escape of muco-pus from the corresponding nostril. In about three quarters of my cases the affection has been limited to one eye. Nevertheless, it is not rare to find from the history that both eyes were attacked to begin with, and that while one has recovered, with or without treatment, the other has failed to do so. Most of the patients who have fallen under my notice appeared to enjoy excellent general health, and I am confident that congenital syphilis did not exist in more than 5 per cent. of the total number—that is to say, in no greater proportion than would be found in poor children attending a hospital for other ailments.

With regard to the frequency of the condition, among 1538 out-patients seen in the ophthalmic department of the North-Eastern Hospital for Children, London, no less than 27—or 1·75 per cent.—were affected. Those figures, however, probably understate the frequency of the ailment, since there are grounds for believing that many cases get well of their own accord without treatment at hospital or elsewhere.

If the secretion expressed from the lacrymal sac be examined by means of cover-glass preparations and cultures, it will be found to contain bacteria. I have conducted many such investigations, and, without wearying you with figures, will merely state that the micro-organisms most commonly associated with the lacrymal mischief are three in number—namely, the pneumococcus, the xerosis bacillus, and the staphylococcus pyogenes albus. Since the two last-named are common inhabitants of the normal conjunctival sac, their presence is hardly likely to be very important.

The researches of modern embryology have taught us that the lacrymal sac and duct are formed from a solid rod, which originates from proliferation of the epithelium at the bottom of the lacrymal groove. By liquefaction of its contents this rod eventually becomes hollow and converted into a canal lined with epithelium. The canaliculi are produced by a bifurcation of the epithelial ridge at the inner canthus. Now, if from any cause delay occurs in the elaboration of the passage, a baby may come into the world with the lumen of the lacrymal tube blocked in various ways. In some instances it is probable that the obstructing medium may be merely inspissated material, while in others it is certain that it may be in the nature of one or more mucous folds. The researches of Rochon-Duvigneaud and others have shown that the lower end of the nasal duct is directed towards the mesial line, and that the canal straightens itself as the superior maxilla develops in height. The canal is divided into compartments by partitions placed at different levels. One of the most conspicuous of these diaphragms forms a reduplicated fold at the lower end of the canal. This is often imperforate, so that the epithelial *débris* filling the passage cannot escape, and may indeed cause the membrane to become distended bladder-wise, a condition described by Bochdalek many years ago under the name *Endblase*. Under such conditions it needs only septic infection to produce the clinical picture of dacryocystitis. The sources of sepsis are, of course, many in number, and may be maternal or otherwise. When the child is born with a discharging lacrymal sac, infection has almost certainly been derived from the maternal passages, and, in my experience, a history of secretion from the genitalia is often forthcoming in such cases. On the other hand, when symptoms are not noticed for several days after birth, the source of infection is likely to be external —for example, from the baby himself or from those about him.

The condition described may be likened to the common and well-known condition of mastitis in babies, where an abscess of the breast may arise from penetration of pyogenic organisms into the epithelial remains that block the ducts of the gland.

The treatment of the condition is as simple as it is satisfactory. Anything like rough handling should be avoided, but careful digital pressure should be made twice a day over the lacrymal sac, with the twofold object of squeezing out secretion through the puncta and of breaking down any obstruction that may exist in the nasal duct. When definite distension of the sac is present, as in the following case, one application of pressure-massage may suffice to cure :

(3) A baby, aged one month, was seen by me on January the 3rd, 1900. At birth the

left eye was noticed to be affected and to have a lump the size of a pea at its inner side. On firm pressure the swelling dispersed suddenly, its contents passing presumably into the naso-pharynx. The child died four days after she was seen by me from "bronchitis and marasmus," but the eye gave no further trouble after the lump had been squeezed.

When compression fails, as it may do when entrusted to the hands of an ignorant or careless mother, we must clear away any obstruction by passing a small probe down the nasal duct. Formerly I was in the habit of slitting the lower canaliculus before probing, but I now content myself with dilating the punctum and canaliculus with a fine, conical instrument, set in a handle, which serves not only to stretch the parts, but also acts as an efficient probe for the duct. As a rule, a single operation of this kind is enough to cure the disease. The nasal duct in babies is extremely short, a point to be borne in mind when using the probe. I have been struck with the fact that in these cases a definite obstruction *qua* obstruction is seldom to be felt. Another point about them is that the end of the probe often feels, as if it were, in a large cavity, doubtless the nasal cavity, which seems much nearer than usual, owing to the shortness of the nasal duct.

There is, I think, a considerable risk of setting up a fistula if the more marked lacrymal collections be lanced from the cutaneous aspect. It is, indeed, a question how many of the lacrymal fistulæ seen in older subjects owe their origin to this method of treating the lacrymal abscess of newly-born children.

(*b*) Lacrymal disease, especially lacrymal abscess, in older children is practically always due, in my experience, either to syphilis or to tubercle. The cases, particularly the syphilitic ones, often recover under constitutional treatment, combined with the simplest local measures, a fact that has rendered me more chary than I once was about interfering surgically. The evidences of syphilis are sometimes none too easy to obtain, although the *café au lait* complexion, more or less depression of the bridge of the nose, and perioral or perianal cicatrices often accompany the disease. In the following case the evidence of syphilis was obtained in a rather curious coincidental way :

(4) Joseph W—, aged 6 years, first seen on January the 6th, 1905, with the right lacrymal sac distended with muco-pus, which appeared in the conjunctival sinus when pressure was made over the prominence. Enlarged glands in neck. The permanent teeth had not yet appeared. The face and complexion, however, rather suggested syphilis. One miscarriage; no still-births. A fortnight later, the patient's left eye showed signs of commencing interstitial keratitis, and later still the same disease developed in the second eye.

The tuberculous nature of a lacrymal affection may be inferred from the family history and co-existing signs of tuberculosis, especially indolent enlargement of the preauricular and neighbouring glands, and can be demonstrated by the discovery of the tubercle bacillus in discharge or scrapings from the affected sac. The local condition of the sac, too, often affords strong presumptive evidence in favour of its tuberculous character. It is apt to show one or more fistulæ, with violaceous and everted edges, and to feel not only thickened but also "lumpy," as if it contained so many polypi. Persistence in an almost unchanged form for months together is also a characteristic of these cases.

Tubercle of the lacrymal sac, despite the comparatively small number of cases reported in the literature, is not, in my experience, an exceedingly rare disease in children.

Congenital Ophthalmia Neonatorum.

It would be a truism to say that serious purulent inflammation of the eyes of newly-born babies is almost invariably due to infection by the specific microbe of gonorrhœa. The bacteriological investigation of 1377 cases of ophthalmia neonatorum in the collective practice of seventeen observers showed that gonococci were present in 61·68 per cent. Among 121 cases of my own, the proportion was 58·67 per cent. We may therefore conclude that roughly two-thirds of all cases of ophthalmia neonatorum are due to the gonococcus. According to figures I have published elsewhere ('Trans. Obstetrical Society of London,' vol. xlv), 80·48 per cent. of the gonorrhœal cases manifest themselves by the appearance of inflammatory symptoms within four days after birth. The actual infection of the baby's eyes with gonococci may come about in three possible ways—(1) in the maternal passages either before or during the act of birth, (2) almost immediately after birth, and (3) one or several days after birth. The second is admitted to be by far the commonest mode of infection. The germ-laden discharge from the maternal passages clings about the eyelids and eyelashes, and, as a rule, is carried into the conjunctival sac, either by the blinking of the baby or by the water, sponges, or towels used for the first bath.

The term "congenital ophthalmia neonatorum," or "*ante-partum* conjunctivitis," has been applied to cases where the baby actually comes into the world with diseased eyes or else develops ophthalmia within a few hours after birth. These cases (of which the existence

can scarcely be doubted nowadays) are interesting from several points of view. They open up certain points as to intra-uterine infections, somewhat of a burning question at the present time in view of the causation of congenital conditions such as opacities of the cornea and anterior staphyloma, and, for that matter, of many other so-called "congenital anomalies" of the eyes.

A child, as is well known, normally traverses the vagina with closed eyelids, and as the junction between the lids is watertight, morbid secretion can hardly enter the conjunctival sac under ordinary circumstances. That could, however, come about during face presentations, the application of forceps, or during digital examinations on the part of the accoucheur. Some of the cases of ante-partum infection can be reasonably explained by a rupture of the membranes having taken place several hours, or even days, before the child was born, thereby allowing gonococci to reach the conjunctival sinuses. The temperature of the parts and the condition of the conjunctiva may possibly account for the relatively rapid development of the inflammatory process observed in some of these cases. Magnus (1) and Bellouard (2) have each reported instances of this kind. Parichew (3) has related a cognate case, where an ill-nourished baby, as soon as it came into the world, was found to be suffering from ophthalmia, together with opacity of the cornea. Gonococci were demonstrated in discharge from the eyes. The membranes had ruptured three days before the beginning of labour, which had lasted about twelve and a half hours and was normal in every way. Of several similar cases I have myself seen I will relate very briefly the leading facts of one :

(5) Thomas W—, aged 3 weeks, was seen on February the 23rd, 1898, suffering from bilateral ophthalmia with gonococci. The baby was born at term after a labour lasting twenty-six and a half hours, and the "waters burst" at least twenty hours before delivery. The right eye was observed to be inflamed as soon as the child came into the world, and the left eye became infected on the tenth day.

In another class of case it is probable that the eyes become infected *in utero*, presumably by the passage of gonococci through the intact membranes or, as Bellouard has suggested, through a lateral rupture of the membranes so trifling as to permit little liquor amnii to escape, although sufficiently large to allow gonococci to penetrate. The first authentic cases of the kind known to me were reported in 1858 by Rivaud-Landrau (4), and since then cases have been published by Chancon, Friedenwald (5), Bellouard (2), Strzeminski, and Armaignac (6). Armaignac's case is so conclusive as to deserve a word of passing mention. An ill-developed infant, weighing only

1786 grm. (3½ lb.), was born at the eighth month, after a labour lasting one and a half hours. At birth the lids were found to be tense, red, and swollen, and upon separating them pus escaped from the eyes. The corneæ were white and macerated. A purulent vulvitis was also present. The mother had suffered from metritis. Nieden's case (7), where a baby born in a caul developed ophthalmia twenty-four hours later, probably belongs to this class. I have heard of a case, but have been quite unable to obtain particulars, where a baby delivered by Cæsarean section was found to present ophthalmia.

The following case has recently fallen under my personal notice:

(6) A baby girl, aged 1 month, was brought to the Evelina Hospital on June the 20th, 1905, on account of double ophthalmia, associated with numerous gonococci in the discharge from the eyes. Both corneæ were hazy, but free from actual ulceration. She was a miserable-looking, "snuffling" baby, weighing 3 lb. 8 oz., with a shock of dark-brown hair (the so-called "syphilitic wig"), and sores upon the anus and vulva. According to the history given by the mother, and afterwards confirmed by the medical man who confined the woman, the baby was born at the end of the eighth month of pregnancy, without instruments, after a natural labour lasting thirteen and a half hours. The "waters burst" seven and a half hours before the completion of the labour, and the only vaginal examination was made by the doctor half an hour before the baby was born. The child was born with red lids and discharging eyes, but the corneæ were clear. Vulvitis was not present. A former baby had suffered from ophthalmia neonatorum.

The evidence, in my opinion, is enough to justify us in believing that, under rare circumstances, there may exist a specific metritis or endometritis, with the passage, direct or indirect, of gonococci into the amniotic fluid. For that matter, the causative microbe need not necessarily be the gonococcus, for I have notes of several cases of congenital ophthalmia where the gonococcus was not present, or at all events could not be demonstrated. Rare cases of congenital anterior staphyloma, of which several have been reported recently, certainly suggest infection, ulceration, and perforation of the cornea at some period considerably prior to birth.

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2. Bellouard.—'Thèse de Paris,' 1892.
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4. Rivaud-Landrau.—'Annales d'Oculistique,' T. xxxvii, p. 66.
5. Friedenwald.—'Medical News,' 1895, p. 257.
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Keratomalacia.

That the cornea may become gangrenous in babies suffering from inanition is a fact that must have struck attentive observers since the earliest dawn of scientific medicine. The condition, indeed, appears to be tolerably common in several parts of the world, as, for example, in Russia, Brazil, Germany, and Japan. A similar condition is found, though rarely, in England, where it affects exclusively babies whose nutrition is at a dangerously low ebb. The disease was described in 1827 by Dr. Joseph Brown, of Sunderland, in a brief communication that bore the significant title of "A Case of Ulcerated Cornea, from Inanition" ('Edin. Journ. of Med. Science,' vol. iii, p. 218). Among the other British authors who have published instances of the disease are Mackenzie, Bowman, Spicer, J. T. Thompson, Ormond, and myself.

Keratomalacia is essentially a disease of children whose ages range from 4 to 12 or 14 months. At the same time, cases are not unknown in newly-born babies, and there are good reasons for thinking that they have now and then been mistaken for ophthalmia neonatorum with destructive corneal lesions.

The disease in infants was attributed by von Graefe to encephalitis, a view adopted by Hirschberg also. The diffuse infantile encephalitis of Virchow, however, is nowadays regarded as non-existent, and so von Graefe's theory has fallen to the ground. It is significant that keratomalacia hardly ever occurs in breast-fed babies, unless, as in Russia, the mother's health has become impaired by long fasting, a thing scarcely likely to happen in this country. Here, without exception, its victims are marasmic, "little wasters" in the pitifully expressive jargon of a children's hospital. The marasmus may be either symptomatic or idiopathic. The causes of the former include tuberculosis, syphilis, and imperfect dieting, with its sequels in the shape of chronic diarrhœa and vomiting. Of seven fatal cases under my care, tuberculosis existed in four (57·14 per cent.), the diagnosis being made during life in two and after death in two of the children. In two of the cases (20·85 per cent.) infantile syphilis was recognised as present during life. In a single case only (10·42 per cent.) did marasmus—that is, a vice of nutrition only—appear to exist. The figures are small, but so far as they go they indicate that tuberculosis is the chief cause of keratomalacia, a conclusion that I had previously reached on other grounds. One must remember that in infancy there are many points of resemblance

between marasmus, on the one hand, and tuberculosis, on the other. Progressive wasting, pallor, and loss of strength are features common to both affections. Many a case assumed to be one of marasmus has turned out upon the dissecting-room table to be one of generalised tuberculosis. A fatal termination is common in keratomalacia. In my present series of ten cases the mortality amounted to 70 per cent. Age has a good deal to do with the result, whether lethal or otherwise. The older the baby, in fact, the better are the chances of recovery.

As regards the cornea, the symptoms of keratomalacia may be summed up in the three words—infiltration, ulceration, and perforation. There is rarely any swelling, redness, or spasm of the eyelids, neither is the eyeball congested, nor, in short, are there any of the appearances that one would naturally expect to find associated with so severe and destructive an affection of the cornea. The eyes often look curiously dry, as it were, and are more or less insensitive to stimuli. A very characteristic feature of the cases is that the baby scarcely resents examination of the eyes, or at most moans feebly and piteously during the necessary manipulations. A common accompaniment of keratomalacia, first mentioned by von Graefe, is a small, triangular, greasy-looking, white patch, situated in the ocular conjunctiva on one or both sides of the cornea. Exceptionally, as in one of my own cases, this may be replaced by a narrow, slightly raised, glistening line, occupying the position of the limbus conjunctivæ. It is doubtless due, like the much commoner triangular area, to the lodgment and multiplication *in situ* of the saprophytic and ubiquitous xerosis bacillus. No specific micro-organism is found in keratomalacia. The ordinary pyococci are, in my experience, invariably present, and considering the bad nourishment of the patients, they are amply sufficient to account for the destructive corneal process. The disease is usually bilateral.

If the baby's nutrition cannot be bettered, the cornea is likely to perforate, a process usually preceded by localised protrusions of Descemet's membrane, which manifest themselves as so many dark-hued areas peeping through the greyish slough that covers the rest of the cornea. Perforation is followed by escape of some of the contents of the eyeball, and the globe eventually shrivels up into a small, white, globular, sightless mass, lying in the depths of the orbit. The more common event, however, is for the baby to die before there has been time for that change to take place.

Treatment.—The first requirement is to place the baby in hospital, where details of feeding can be attended to properly, and the child

can be kept warm, if necessary in an incubator. The general treatment is that suitable for marasmus. The administration of alcohol, in the form of brandy, has seemed to me to be most helpful. Cod-liver oil has a distinct value in these cases, especially when used as an inunction. Neatsfoot oil is as valuable as cod-liver oil, and is certainly much more economical. If syphilis be present, recourse must be had to mercury. The local treatment of the eyes will include the frequent use of douches of warm boric lotion. The employment of physostigmine has been warmly recommended by Dr. J. Thalberg ('Archives of Ophthalmology,' 1883, p. 211), who has seen much of the disease in Russia. My own experience leads me to rank physostigmine very highly, and it is sometimes surprising to observe how quickly the cornea casts off its sloughs under the influence of that remedy. A solution containing one grain of physostigmine sulphate to the ounce of water should be dropped into the eyes every four to six hours according to the severity of the case. During the more acute stages of disease the eyes should be kept tied up, and it goes almost without saying that the utmost gentleness should be exercised in making the necessary applications, examinations, and so forth.

Buphthalmos.

There are several points about this rare and peculiar malady that are still somewhat of a mystery to ophthalmic surgeons.

Buphthalmos is either present at birth or, less commonly, develops at a very early age. It may affect one or both eyes. Its leading symptoms include : (1) increased intra-ocular tension ; (2) progressive enlargement of the entire eyeball, revealed clinically by the greater dimensions and altered curvature of the cornea, whence the names "ox-eye" and "megalocornea" sometimes applied to the disease ; (3) pathological cupping of the optic disc ; (4) haziness of the cornea ; and (5) defective sight and a degree of photophobia.

It is established that the primary departure from the normal in buphthalmos lies in the increased intra-ocular tension, and that the other symptoms, as enlargement of the eyeball and cupping of the optic disc, are dependent upon this feature. Hence, buphthalmos is nowadays usually regarded as a form of congenital or infantile glaucoma. In progressive cases the iris may become so stretched and attenuated as to be torn, thus producing, as it were, several artificial pupils, as happened in a case of my own.

There are comparatively few cases of buphthalmos where the cornea

retains complete transparency. Indeed, the corneal lesions may now and then, as in the following case, form the prominent feature of the disease :

(7) Edith P—, seen when about 3 months of age, showed a slight degree of buphthalmos. The history was that the baby was born on June the 30th, 1900, and that she had always been remarkable for possessing "a fine pair of eyes." On awaking on the morning of September the 8th, 1900, the right eye was noticed to be cloudy a condition diagnosed by the two medical men who were summoned as due to "keratitis." The eye was not red, but there was evidently some photophobia, since the baby, for the first time in her life, screwed up her eyes when taken out of doors. The

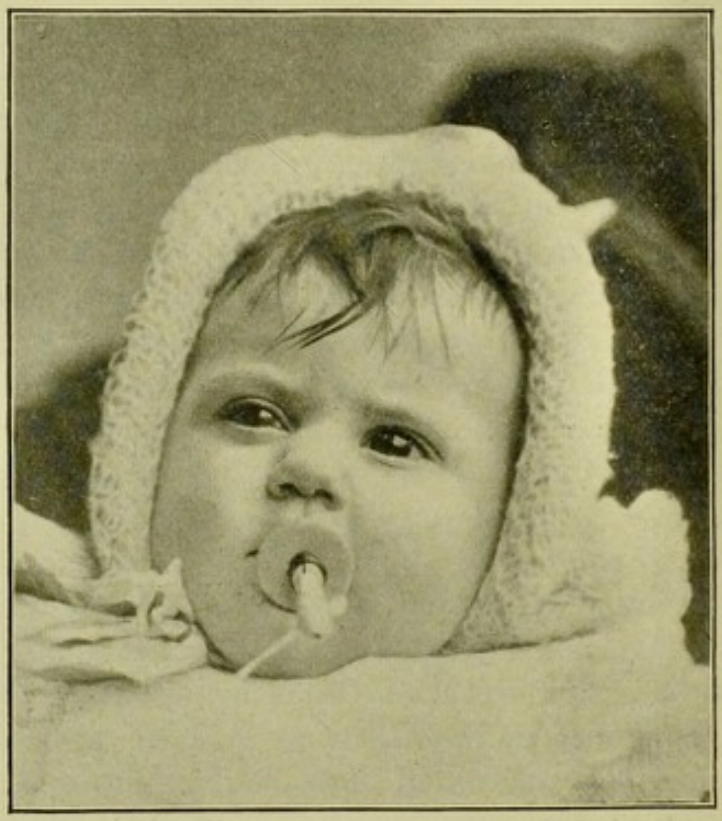

Fig. 1.—Mild buphthalmos.

cornea remained cloudy until September the 11th, when it began to clear, but a few days later (September the 17th) it again got dull. When I saw the baby a few days later, there was slight but definite enlargement of both eyes ; the corneæ were somewhat milky ; tension was *plus*. The baby "snuffled" slightly, and the bridge of the nose, as shown by the photograph, was slightly depressed. The child was, however, well nourished, although very bad tempered. The baby remained under my immediate observation for several weeks, during which time the corneæ alternately cleared and became cloudy, and I satisfied myself that the alterations in transparency were connected with a corresponding change in the intra-ocular tension and presumably with an obstruction to the circulation of lymph. The patient was treated for about three and a half years with physostigmine locally and mercury with chalk internally, and on April the 9th, 1904, the following note was made : "Child distinguishes objects readily with either eye. Tension normal. Right cornea 12 mm. and left cornea 11 mm. The right eye looked larger than the left. There was a good fundus reflex,

but the pupils were so much under the influence of the physostigmine that no satisfactory view of the fundus could be obtained." On March the 21st, 1905, the child was seen again, and a note made to the following effect: "A few small nebulæ mark the site of a recent phlyctenular efflorescence. The child is said to see well at a distance, but she looks close at coloured pictures. Transverse diameter of each cornea 12 mm. The right eye, however, looks larger than the left, owing to the cornea of the former being more globular than that of the latter. Anterior chamber deep, especially on the right side. Tension normal. Discs deeply but not universally cupped. A myopic crescent is present, especially in the right eye."

Pathological examination of eyes with buphthalmos has shown as a constant lesion adhesions between the root of the iris and the periphery of the cornea. My own specimens, derived from three eyes,

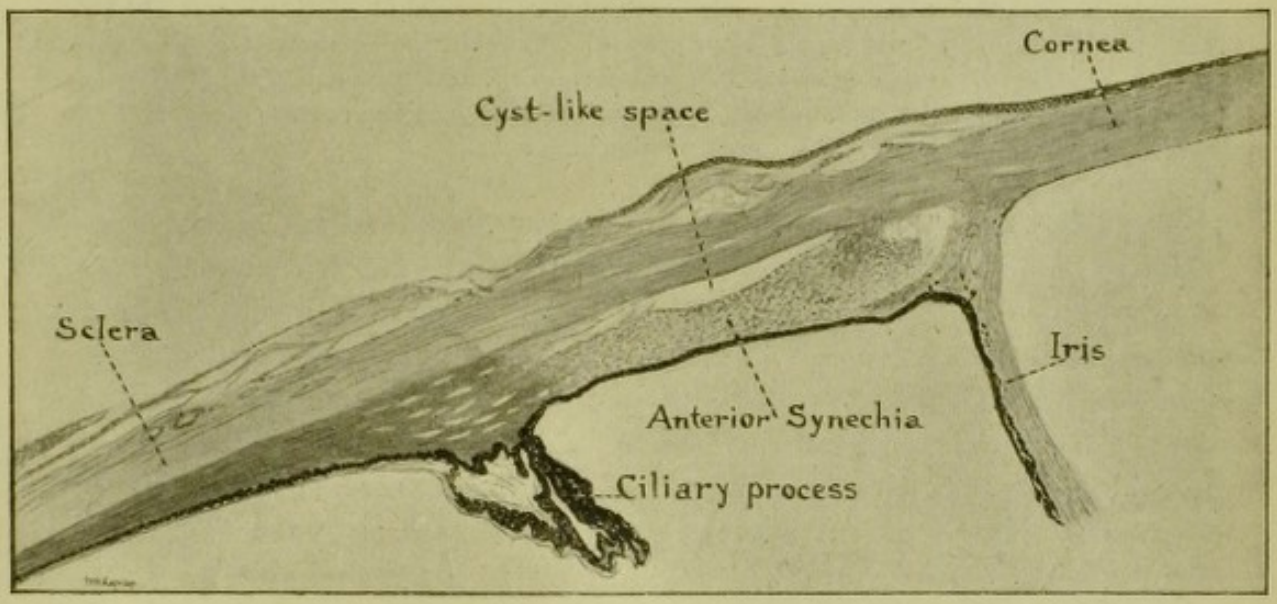

FIG. 2.—Absolute buphthalmos. (Evelyn L—, aged 7 years.) The specimen (magnified 20 diameters) shows that a wide fibro-cellular adhesion exists between the root of the iris and the corresponding part of the cornea. The two structures are not in accurate apposition at all points, so that a cyst-like space exists at one place. The specimen also shows the ciliary processes and the unaffected part of the iris and cornea. It will be noticed that the iris is retroflexed.

all go to prove that this is the outstanding pathological feature of the disease. According to one view, of which Mr. E. T. Collins (1), if not the actual originator, is, at all events, the chief exponent, these anterior peripheral synechiæ—for that is what they amount to—are caused by developmental defects. Mr. Collins supposes that the separation of the iris from the posterior surface of the cornea, which should occur normally prior to birth, fails in whole or part, owing to reasons that are yet imperfectly understood. In this way a congenital anterior synechia, extending more or less around the sinus of the anterior chamber, is formed. According to another view (of which I am personally an adherent), the adhesions in question result

from an intra-uterine inflammation of the iris and adjacent structures. I am myself convinced that this represents the correct view, and I would say that such intra-uterine inflammation is closely connected with congenital syphilis. At all events, of nine patients under my care, seven exhibited conclusive signs of syphilis, while in the remaining two there was strong collateral evidence for believing that disease to exist. I quote the following as a typical case of the kind :

(8) Violet M—, aged 5 weeks, attended the North-Eastern Hospital for Children on February the 9th, 1905, on account of discharging eyes. The baby was breast-fed. There had been two living children in the family, then three born dead at about term, and, lastly, the patient. She was an emaciated, "snorting" infant with depressed bridge of the nose and red and desquamating palms and soles and excoriated skin about the mouth. The corneæ were somewhat globular and slightly hazy, and both eyes were larger than natural. Tension was thought to be a little raised. Under prolonged treatment (physostigmine and mercury with chalk), the baby regained excellent health, tension became normal, and the corneæ clear, and the eyeballs showed no tendency to further enlargement.

Buphthalmos occasionally occurs in several members of the same family. The most remarkable instance of the kind with which I am acquainted was reported in 1898 by Dr. Walter B. Johnson ('Annals of Ophthalmology,' vol. vii, p. 539). The first three children, two boys and a girl, of a "strong and healthy" couple, were not affected. The next three children, all females, manifested symptoms of infantile glaucoma, a disease cured, or at all events held in check, by the performance of sclerotomy or iridectomy at a very early age. At first sight this hereditary tendency to buphthalmos might be thought to tell in favour of Mr. Collins' developmental theory, but it is, to my mind, more in favour of syphilis contracted in the interval between the birth of the non-affected and the affected children. It is perhaps worth noting that of the photographs given of the three patients in Johnson's communication, two distinctly suggest a syphilitic type of face.

Whichever view be adopted as to the origin of the synechiæ, whether congenital or inflammatory, the result of the adhesions as regards the eyeball will, of course, be the same, namely, blocking of the angle of the anterior chamber, and consequently impeded exit of fluid into Fontana's spaces and Schlemm's canal. This would account for the deep anterior chamber which is so marked a feature in most cases of buphthalmos.

There are yet other theories with regard to the pathogenesis of buphthalmos. Thus, Angelucci believes that the condition is essentially connected with disturbance in the vaso-motor mechanism. He explains the disease by assuming a congenital lesion in the bulbar

centre of the sympathetic, causing first vaso-dilatation and then hyper-secretion. In connection with Angelucci's theory, the fact may be mentioned that Lodato discovered a marked hyperæmia of the superior cervical ganglion on the affected side of a rabbit presenting unilateral congenital buphthalmos.

Buphthalmos must be carefully distinguished from an allied condition which very occasionally follows cases of severe interstitial keratitis and gonorrhœal ophthalmia. These may be regarded as instances of secondary buphthalmos, reserving the name "primary" for the disease now under discussion. The diagnosis of primary buphthalmos can offer no difficulties, particularly if one eye alone be affected. It is, however, possible that the slighter grades may be overlooked or mistaken for something else, as, for example, so-called "congenital opacities of the cornea." Again, I have known the disease confused with interstitial keratitis, as in the following case:

(9) Lily W—, aged 6 months, attended the North-Eastern Hospital for Children on October the 24th, 1900, when a diagnosis of interstitial keratitis was made on account of the cloudy corneæ and of a history pointing to syphilis. When I saw the child a week later, I found a "snuffling" infant, who showed traces of a recent ulcerous eruption of the vulva and anus. The eyes were said to have been affected for about three weeks. The eyeballs were obviously large, the transverse diameter of the corneæ measuring 14 mm. The corneæ were globular, with a good deal of haze. The last-named took the form of a Y-shaped figure, whose prongs were occupied by faint, punctate opacities. Discs cupped. Tension raised. The muco-purulent discharge present was connected, I thought, with congenital syphilis.

In buphthalmos, as in most other diseases, we must expect to meet with mild and severe cases, a somewhat obvious point upon which too little stress is sometimes laid by writers on the subject. Treatment, in my opinion, is by no means so hopeless a thing as one might gather. The disease, it is true, may progress to absolute blindness, but, on the other hand, it may undergo cure, either spontaneously or otherwise during the first few years of life. In spontaneous cure it has been suggested that the adhesions shown to exist between the iris and the cornea become stretched coincidently with the growth of the globe, and thereby cease to obstruct the passage of aqueous humour from the eye (E. T. Collins).

Cases of mild buphthalmos are unquestionably amenable to local treatment by myotics and general treatment by mercurials, but, so far as I have seen, treatment, if it is to be crowned with ultimate success, must extend over several years. In several of the more severe cases I have obtained good and apparently permanent results by the performance of large iridectomies, and I am by no means

certain that the ordinary dictum, namely, not to operate, is one that should invariably be acted on. It is clear, however, that operation should be undertaken at as early an age as possible. In long-standing cases the iris is far more likely to be torn away from the synechiæ instead of from the ciliary body, which is necessary for relieving the tension. The analogy with very chronic glaucoma, in which the same thing is liable to occur, is complete. The different views held with regard to the value of operation are possibly connected very closely with the stage of buphthalmos at which iridectomy or sclerotomy is adopted. Professor Haab (2) tells us that "infantile glaucoma can be cured by repeated sclerotomy if begun early enough"; Dr. B. Stölting (3) by the same means has obtained encouraging results in five children whose ages ranged from 7 months to 2 years; Schoenemann (4) has reported seven cases (thirteen eyes) treated with success by non-peripheral iridectomy. Schoenemann's cases were watched for periods ranging from three to six years, so that his communication is particularly valuable. Dr. H. R. Swanzy (5), on the contrary, says in so many words that "iridectomy and sclerotomy are alike followed by disastrous results in this disease." When all is said and done, however, it is probable that truth, as usual, lies somewhere between the extreme positions taken up by the authors quoted.

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DEFORMITIES OF THE SKULL AND DEFECTS IN SIGHT.

There are three malformations of the skull that are apt to be accompanied by defective sight, viz., (1) microcephaly; (2) hydrocephaly; and (3) oxycephaly.

(1) *Microcephaly.*—Abnormal smallness of the cranium is usually a congenital defect in the sense that a baby enters the world with a skull the capacity of which is smaller than normal, while its frontal portion is flat and receding. It used to be thought to be due to premature synostosis of the cartilage between the basi-sphenoid and the basi-occipital bones, but of late years considerable doubt has been cast upon that view. The children are commonly idiotic, and they present the conditions often associated with mental deficiency,

such as a vacant expression, deafness, and backwardness in walking and talking. The condition is apt to be reproduced in other members of the family.

It is not uncommon for a microcephalic infant at the age of six or eight months to be brought with the statement that he cannot see. This idea is sometimes well founded, and in that event examination of the fundus allows one to distinguish between two groups of cases: (*a*) the fundus appears to be healthy, and the defective sight to be merely a part of the general deficiency from which the child suffers, and (*b*) the fundus is found to include areas and patches of choroiditis, having no particular shape and occupying no particular position. The existence of the last-named class (*b*) has been noted by Mr. E. Nettleship ('Royal London Ophthalmic Hospital Reports,' vol. xi, p. 356), who in 1887 reported four cases of the kind. Mr. Nettleship thought that there was no evidence of syphilis, a view with which I am in accord. Personally, I have never been able to convince myself that the choroidal changes, which are probably of intra-uterine origin, have anything to do with the blindness, which I consider is amply accounted for by the mental condition of these children. In other words, in this class of case I am inclined to regard the fundus changes as merely coincidental.

(2) *Hydrocephaly.*—The pathogeny of defective sight or actual blindness not seldom met with in cases of chronic internal hydrocephalus is, of course, well understood. It was known to the older writers under the name of "hydrocephalic amaurosis." It is due to simple optic atrophy, resulting from the pressure of the distended third ventricle upon the optic tracts and chiasma at the base of the brain. The following is a typical case:

(10) Jessie S—, aged 3 years, seen at the Evelina Hospital on January the 18th, 1904, suffering from hydrocephalus, stated to have commenced at 15 months of age. Circumference of head, 23½ inches. Child quite blind, taking no notice of the flash of a mirror and not flinching when the finger was thrust forwards towards the eyes. Retinal vessels of good size. Both optic discs showed a large pallid cup, and surrounding this the papilla was of a greyish-white colour. No squint.

The optic atrophy may be incomplete, and, as in the case to be next quoted, associated with paresis of an ocular muscle:

(11) Beatrice P—, aged 1 year and 7 months, seen at the Evelina Hospital on March the 16th, 1900. According to the history there had been a squint and the head had been getting bigger for six months. No convulsions. "Water on the brain" had been diagnosed by my colleague, Dr. Soltau Fenwick. Child almost but not quite blind. Abduction of eyeballs most imperfectly carried out, and a convergent squint present. Both optic discs were greyish, particularly as regards the inner side.

I have myself once only seen optic atrophy in hydrocephalus preceded by optic papillitis :

(12) Daniel D—, aged 1 year and 9 months, seen at the North-Eastern Hospital on March the 11th, 1896, suffering from a rather marked degree of hydrocephalus. There were central choroidal changes in each fundus, especially in that of the left eye. The optic discs were swollen, hazy, and of whitish-grey colour. The discs afterwards became quite pallid, so that from mere inspection it would have been impossible to distinguish them from a condition of so-called simple atrophy.

Depression of the eyes and nystagmus are common features in extreme cases of hydrocephalus.

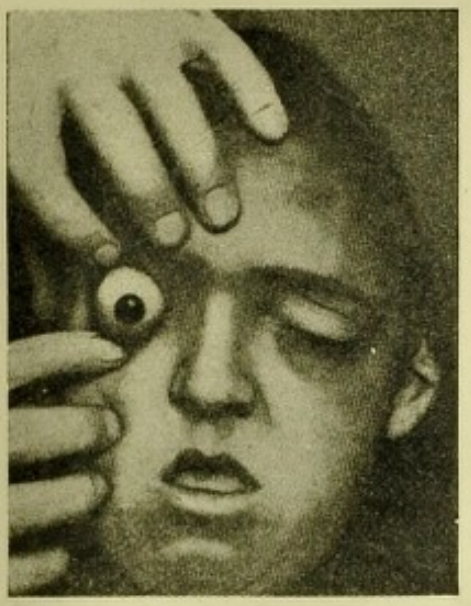
(3) *Oxycephaly.*—This curious deformity (known in France as *crane en forme de tour* and in Germany as *Turmschädel*) is charac-

Fig. 3.

terised by abnormal height of the anterior portion of the skull, so that the frontal region is capped by a bony prominence and assumes a steeple-like form. Hence the name "tower skull." Virchow's view was that the deformity was due to premature ossification of the sutures between the parietal bones and the occipital and temporal bones, resulting in a kind of compensatory development involving the anterior portion of the cranium. The condition, as a rule, is congenital. In my experience, extreme instances of the deformity are exceedingly uncommon. Although the patient may be mentally defective, yet he is usually intelligent, probably because the brain accommodates itself to the changes in the contour of the cranium. Owing to the shallowness of the orbits, the eyes become very prominent ; so much so indeed that they may be partially extruded from

the orbit by slight digital pressure. Donaldson (3) has related a case where exophthalmos was so pronounced that on several occasions one eye became partially dislocated and had to be replaced by the patient's father. The accompanying illustrations (taken from a recent communication by Professor Uhthoff [9]) represent a weak-minded lad, aged 15 years, the subject of oxycephaly, whose right eye was so exceedingly prominent that it could by slight pressure be dislocated in front of the eyelids. Divergent squint and nystagmus are common in these cases, and paralysis of ocular muscles is not unknown. But their chief ophthalmological interest centres around the fact that a certain number of the patients are more or less blind, in consequence of optic atrophy, of post-neuritic type, beginning in comparatively early life. It is significant that in the three examina-

Fig. 4.



tions after death of patients who suffered during life from oxycephaly and papillitis constriction of the nerve in the optic foramen was found (Enslin). At the same time, it has been suggested that the papillitis may result from augmentation of the intra-cranial pressure or from meningitis. Another school teaches that the deformity of the skull and the inflammation of the optic nerve result from a common cause, the nature of which is at present unknown. Oxycephaly may affect more than one member of the same family, as in cases described by George Carpenter (1) and Velhagen (6). Carpenter's cases were in two sisters, aged about 2 and 4 years respectively, who, in addition to marked oxycephaly, presented a perfect arsenal of associated defects. Indeed, the condition often co-exists with other stigmata of degeneration, such as hallux varus, genu valgum,

highly arched palate, polydactilism, webbed fingers and toes, meningocele, hernia, deformities of the ear, and so forth.

The following case of oxycephaly fell under my notice recently:

(13) Albert R, aged 17 years, examined April the 11th, 1904. The lad, although he had never attended school, had nevertheless managed to teach himself how to read. He spoke well, and was distinctly intelligent. He was not deaf. His height was 4 ft. 11½ in., and his weight 5 st. 7 lb. 8 oz. A general examination disclosed the existence of the following defects or malformations, viz. a highly arched palate, alar scapulæ, deep post-anal dimple, and a trace of flat-foot. The lad's head was small

Fig. 5.

and of the oxycephalic type (*see* photographs). The high, keel-like forehead terminated in a definite osseous lump situated at the junction of the coronal and sagittal sutures —*i. e.* at the site of the anterior fontanelle. Measurements of the skull were as under: (*a*) Horizontal circumference, 49 cm.; (*b*) over vertex, from ear to ear, 31 cm.; (*c*) arc from root of nose to occipital protuberance, 35 cm.; (*d*) orbital height 4 cm.; and (*e*) orbital width, 5·5 cm. The nose was deflected strongly towards the right. The eyeballs were very prominent, but on an effort being made, could still be covered by the lids. The prominence of the eyeballs was such that the corneæ projected beyond a straight edge applied to the supra-orbital ridge above and to the junction of the malar and superior maxillary bone below. There was no pulsation, and the eyeballs could not be pushed back into the orbits. The finger could be readily

passed above between the globe and the roof of the orbit and be passed back some distance beyond the equator of the globe, which under those circumstances became pushed markedly forwards. The left eyeball was divergent. There was no obvious nystagmus. R. V. $= \frac{6}{60}$. L. V. $= < \frac{6}{60}$. Mixed astigmatism in each eye. R. E. the optic disc was greyish-white, with opaque nerve-fibres at its upper and lower border. It was oval down and out, and its vessels were markedly irregular in position and distribution. L. E. pronounced post-neuritic atrophy. The vessels, which were of good size, were bundled together upon the inner third of the disc, where they formed a contorted and convoluted mass. The left disc was much whiter than that of the other eye. It is to be noted that although coarse nystagmus did not exist, yet during ophthalmoscopic examination the eyes manifested slight swaying movements.

Fig. 6.

The family history of Albert R— was instructive. In addition to two miscarriages, there had been five children, of whom three, including the patient, survived. Albert was the first-born. Next came a baby girl, who died at 15 months from convulsions. Her head was deformed, and her eyes became prominent two months after birth. Two boys, both still living, came next, but they showed no deformities. Finally, a girl, who died from "convulsions and teething" at 6 months, and who was not deformed in any way, was born. The mother of the patient was examined, and found to have shallow orbits, retreating forehead, and prominent eyes.

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Nystagmus in Children.

In children, as in grown-up persons, the curious involuntary oscillations of the eyeball known as nystagmus may be due to very different causes. In childhood these include any factor capable of interfering with the development of the faculty of sight and of fixation, such as ophthalmia neonatorum, congenital cataract, corneal opacities, very high myopia, or choroido-retinitis, due to syphilis or other disease.

I desire, however, more particularly to discuss three causes of nystagmus that collectively are commoner in children than all the other causes put together. These are: (1) Gyrospasm; (2) Partial albinism; and (3) Post-meningitic or hydrocephalic nystagmus.

(1) *Gyrospasm.*—It is not uncommon to have a baby brought on account of the more or less recent development of quivering, or rolling, or twitching, or "twittering" movements of the eyes. The history of the case, together with the peculiarities of the nystagmus, cannot fail to strike an attentive observer. As regards the nystagmus, the condition is often unilateral, or it is more pronounced in one eye than in the other. If both eyes are affected, one eye may move up and down and the other in and out, whereas in the common run of cases, due to other causes, nystagmus is bilateral, and occurs in a definite direction in both eyes, so that they retain their relative parallelism. In another very peculiar form, which has been named by Dr. John Thomson "convergent nystagmus" ('British Medical Journal,' March 30th, 1901), the eyes become convergent in successive jerks, and then become divergent in the same sort of way. The nystagmus may be associated with or replaced by a marked retraction of the upper eyelids, giving the face a peculiar staring appearance that once seen is not readily forgotten.

(14) Richard H—, aged 16 months, seen in February, 1896. He was affected with lateral nystagmus, occasional convergence of the right eye, and marked retraction of each upper lid, varying in extent during the time the child remained under observation. Optic disc, retinal vessels, and fundus generally normal. Slight rickets was present.

In these children you will often notice jerking, rolling, or nodding movements of the head, although it is well to remember that nystagmus may precede the latter by several days or even weeks.

(15) Florence M—, aged 8 months, brought on February the 24th, 1897, because the right eye had begun to "twitch" five days previously. She had always been a "nervous" child, frightened of falling, and startled by a sudden noise. Mother and father stated to be "shocking nervous." No history of fits, chorea, or mental derangement in the family. Fifth child. Breast-fed exclusively. Upon examination, lateral nystagmus, much more marked in right eye. No hippus. No movements of head. Fundi normal. Baby notices light. March the 10th, 1897: During the last week baby has been observed two or three times a day to nod her head and to move it from side to side. She is less nervous, and the eye movements are not so pronounced as they were. March the 24th, 1897: Eyes distinctly better, but movements of head more marked.

Even under those circumstances, the recent onset, the peculiarities of the nystagmus, and the age of the child (usually from 6 to 18 months) should warn you that you are dealing with an instance of what is called "gyrospasm," "head nodding," or "spasmus nutans." These children show signs of rickets (often slight), and it may be surmised with a fair show of reason that the associated intestinal derangement generates poisonous substances, which, absorbed by the blood, are enough to throw out of gear, as it were, the recently-acquired and presumably still feeble movements of co-ordination. Not the least curious point about this curious neurosis of co-ordination is that cases are commoner in winter than at other seasons of the year, a fact supposed to be in some way connected with the absence of sunshine.* This observation had led to an analogy between gyrospasm, on the one hand, and miners' nystagmus, on the other. It is doubtful how far the disease is connected with dentition. The subjects of spasmus nutans are often unusually intelligent and very good tempered, although they are apt to be startled by any sudden noise. Temporary lapses into unconsciousness may be present, as in the case to be next related:

(16) Beny S—, aged 9 months, seen at the Evelina Hospital on November the 29th, 1901. Child never well. At 7 months pneumonia and bronchitis. Always more or less "convulsed." Breast-fed. Eyes said to have "wobbled" since he was born. Child notices sometimes, but not at others, when he is stated to "look vacant." Upon examination, lateral nystagmus. Pupils equal and active. Child "snuffles" vigorously. He follows a bright object held before his eyes imperfectly. For the last

* Schapringer agrees with Randnitz in thinking that the main cause of the nystagmus is to be found in keeping babies in comparatively dark rooms in which there are one or several spots of bright light, the fixing of which exhausts the retina ('Centralbl. für prak. Augen.,' August, 1905).

six weeks his head has been rolling from side to side. Fundus reflex good, and no changes can be made out in the optic discs or central region. Child starts and screams at sudden noises.

There is often a history of gyrospasm in the baby's brothers or sisters. The following is an excellent example of this point:

(17) William B—, aged 2 years, seen on February the 13th, 1901, on account of an ulcer of the left cornea. The child could neither walk nor talk. Cranial bosses. No facial irritability (Chorstek's sign). When five weeks old the child had developed lateral movements of the head and eyes. The former passed away, but horizontal nystagmus still exists. Rickets. There had been six other children in the family, and all of them rolled their heads and eyes from side to side at the age of 6 or 7 weeks. These symptoms persisted during the period of dentition and then disappeared.

After lasting for a few months, the disease always gets well, with or without treatment, and the symptom that first occurred, be it nystagmus or head movements, is, in my experience, generally the first to disappear.

The best drug to give is antipyrin, in one-grain doses two to four times a day. The bromides have also been recommended.

(2) *Partial albinism.*—The light hair and eyelashes, the glowing pupils, the pink iris, the nystagmus, the photophobia, and the defective sight of a typical albino make up a picture that, once seen, can never be forgotten. Mr. E. Nettleship, however, under the name of "partial albinism" has recently directed attention to an interesting group of cases in whom imperfect sight and nystagmus coincide with much slighter signs of albinism ('Royal London Ophthalmic Hospital Reports,' May, 1902). The condition is apt to run in families and to affect the males, although the tendency, as in hereditary optic atrophy and in several other conditions, is transmitted downwards by the female members. Mr. Nettleship's observations furnished me with a clue to the explanation of a form of nystagmus in children with which I had been familiar for some time. The condition, in my experience, is not uncommon, although its essential nature is often overlooked or misinterpreted. The following case, seen only the other day, is tolerably typical of the condition:

(18) Henry G—, aged 6 months, examined on May the 26th, 1905. About one month after birth his eyes were noticed to "wobble." He was accordingly taken for advice to the special department of a London general hospital, where the surgeon is reported to have told the mother that "the child never had seen and never could see." Upon examination, I found a baby in good health, and with normally developed skull, who showed lateral nystagmus, but who responded readily to the flash test. Much compound hypermetropic astigmatism existed. The choroidal vessels were unusually distinct, except in the yellow spot region of the fundus oculi. The hair, eyebrows,

and eyelashes, although not actually white, were light-coloured, and the iris contained a certain amount of pigment, having a bluish tinge. Henry was the fifth child in the family. The other children were said to be of very light complexion. The eldest, indeed, was stated to have white hair, bad sight, and spectacles.

In the following cases three children belonging to the same family were the subject of this interesting condition :

(19, 20, and 21) On February the 25th, 1903, William L—, aged 6 years, was brought to the North-Eastern Hospital with the complaint that his sight was bad and that his eyes "stuck together" in the mornings. There was slight chronic palpebral conjunctivitis, with the Morax-Axenfeld diplobacillus in the scanty mucus secretion. Lateral nystagmus existed. 3·5 D. of hypermetropia. The lad had reddish hair, eyebrows, and eyelashes, and his iris was grey-blue. The choroidal vessels were visible all over each fundus, except in the yellow spot region. On inquiry it was ascertained that William L— had two sisters, whose eyeballs had always been unsteady. The younger of these two girls, aged 10 years, presented nystagmus and shaking movements of the head, light hair, blue irides, pigmented cilia, and an albinotic type of fundus oculi. 2·5 D. of hypermetropia in the right eye, and in the left the same, with 1 D. of astigmatism, axis vertical. The elder sister, aged 11 years, manifested very similar conditions, general and ocular.

(3) *Post-meningitic and hydrocephalic nystagmus.*—When speaking of amblyopia in children I shall describe a fleeting form associated with meningitis. In certain cases these children exhibit a form of nystagmus, which persists for several years and, for aught I know to the contrary, permanently. This form offers no special peculiarities, but its nature can usually be recognised from the history of the case. The nystagmus associated with cases of severe hydrocephalus has already been mentioned earlier in this Lecture.

Amaurosis in Children.

Leaving on one side hysterical amblyopia, night-blindness, amaurosis partialis fugax, congenital amblyopia, and amblyopia from non-use, none of which is common in children except the two last-named, there remain for description three conditions that occur exclusively in quite young children, viz., (1) Acute cerebral amaurosis; (2) Post-convulsive amaurosis; and (3) Amaurosis following prolonged blepharospasm.

(1) *Acute cerebral amaurosis.*—That many infants suffering from meningitis lose their sight from papillitis followed by optic atrophy, is a fact known to every student of medicine and duly set forth in every text-book. There exists, however, a type of case which is less common, where during the course of a cerebral attack sight is altogether lost without ophthalmoscopic signs of papillitis or atrophy, and the particular interest of such cases lies in the fact that the

children often recover both sight and health at a later period. I am convinced that this "fleeting amaurosis," as I have elsewhere ventured to call it, is not so generally recognised as it should be, an omission the more curious seeing that its essential features have been worked out by Mr. Edward Nettleship ('Ophth. Soc. Trans.,' vol. iv, 1884) and by Dr. William Gay ('Royal London Ophthalmic Hospital Reports,' vol. xiii, 1893, p. 404), who have published excellent descriptive papers on the subject.

In my own experience, the patients are usually below two years of age. There is a history of recent illness marked by retraction of the head, fits, twitchings, drowsiness, stupor, rigidity, vomiting, bulging of the anterior fontanelle, headache, and perhaps other symptoms suggestive of an inflammation of the meninges of the brain. The temperature may be of distinctly febrile type. Obstinate constipation is sometimes present. At some period during the illness, which runs a chronic course, the mother finds that her baby ceases to take notice of things and to recognise familiar and, it may be, favourite objects. The loss of sight appears to be complete, and, so far as can be made out, to come on almost suddenly. At this stage the pupils, in my experience, are larger than normal, and possess little, if any, direct response to light, but beyond that I have never succeeded in obtaining any objective evidence of organic changes in the eyes. In the further course of the affection cerebral symptoms pass away, the child slowly regains his health, and sight returns gradually. The pupils react to light, and ophthalmoscopic appearances are negative.

The following case is typical:

(22) Henry K—, aged 7 weeks, attended the Evelina Hospital on July the 26th, 1901, with the statement that he had lately lost his sight. The baby, who was breast-fed, had suffered from vomiting for three weeks and from diarrhœa for a few days. His head was markedly retracted. The pupils were equal and active; there was no squint; the optic disc and retinal vessels showed no changes. The baby appeared to be blind, inasmuch as he took no notice of a bright light. August the 2nd, 1901—The child had several "convulsion fits" soon after leaving the hospital, a week ago, and these have recurred in the interval. He still vomits. August the 9th, 1901—The head is still retracted, but the child is better in himself, and the vomiting has almost stopped. He notices nothing; there are no ophthalmoscopic changes. August the 23rd, 1901—Neck no longer stiff. Baby putting on flesh. Pupils equal, of medium size, but with little reaction to light. The child recognises his mother when she comes into the room. September the 20th, 1901—The child now "coos" when his mother comes into the room, and notices things about him. His sight is stated to improve daily. Pupils equal, with better action to light. Fundi normal. General health now satisfactory.

That a simple basal meningitis existed in most of my cases was

the opinion held both by myself and by the physicians in more direct charge of the patients.

Such, in brief, are the essential features of what Dr. Gay has called the "acute cerebral amaurosis of infancy," than which it would, I venture to think, be hard to find a better descriptive name.

Nothing certain is known as to the anatomical basis for the blindness. Elsewhere ('Reports of the Society for the Study of Disease in Children,' vol. ii, 1902, p. 276) I have ventured to suggest that the essential factor is closure of the foramen of Magendie, leading, in the first place, to distension of the third ventricle, and then to pressure upon the optic chiasma. If from any cause the adhesions yield early enough, the consequences would appear to be twofold—first, relief to the meningitic symptoms, and, secondly, restoration of vision before the onset of atrophic changes in the optic nerve. Another view is that the blindness results from a temporary inhibition of the visual centres by toxic meningeal products (James Taylor).

(2) *Post-convulsive amaurosis.*—Not long ago Dr. Henry Ashby and myself described ('Reports of the Society for the Study of Disease in Children,' vol. iii, 1903, p. 197) a series of five cases, where temporary blindness seemed to be the direct outcome of severe convulsions in young children. The common history was that the infant had been convulsed for several hours or days, and that when he regained consciousness, he was blind and his arm and leg were paralysed. The sight returned completely, but not the power in the limbs. In other cases paralysis had been but slight, and the paresis disappeared before the amaurosis.

It may be objected that convulsions do not constitute a morbid entity, but are merely accidental complications of many pathological states. This is doubtless true. Our point was that the convulsions, whatever their origin, were themselves the actual cause of the blindness. The "explosions," or nerve-storms, may involve the visual cortical centres as well as the Rolandic motor areas, or the speech centre, and the period of discharge be followed by one of exhaustion. In those cases associated with hemiplegia, which becomes permanent, there can be little doubt that there is a grave organic lesion, such as thrombosis or hæmorrhage, involving part of the Rolandic area or its neighbourhood, but whether the lesion is the cause or the consequence of the convulsions in any given case must, for the present, remain an open question. The stage of coma, which is often prolonged for some time after the convulsions have ceased,

may be the result of exhaustion or of toxin poisoning, and, when consciousness returns, the visual centres are left in an anæsthetic state, or the motor speech centre, or other motor centres, may remain in a temporarily exhausted condition.

The following case, which has recently fallen under my notice, is tolerably typical of the class described by Dr. Ashby and myself:

(23) Mabel B—, aged 2½ years, was seen by me on August the 15th, 1904. *History*—The patient was born on November the 5th, 1901. After suffering from thrush and from whooping-cough, she began to waste at about the end of the fifth month. When the illness had lasted two months—*i. e.*, at the end of the seventh month of life—her mouth, eyes, hands, and legs became convulsed, a condition that lasted for one week. For a month the baby "lay like one dead." She then began to recover, and it was found that she was blind. The amaurosis persisted for about two months, and then slowly passed away. The first sign of returning sight was that when the baby was taken into the light she shut her eyes, a thing she had never done before. Next, she smiled and held out her hands on seeing her mother; food was then noticed. A further stage was marked by the baby, on hearing anybody enter the room, looking about her to see who it was, and by "fixing" the newcomer for a moment. The mother thinks that, to begin with, sight could be sustained for a few moments only. Upon examination, the child was well nourished, walking, and talking well. She readily noticed things held before her, and, baby-like, grabbed at them. Pupils equal and active. Disc, vessels, and central region of each fundus normal. Slight nystagmus on lateral movements of the eyes.

The conclusions drawn by Dr. Ashby and myself regarding this form of amaurosis were as follows: (1) that there is a form of amaurosis which occurs in infants or young children which is post-eclampsic, due to anæsthesia of the visual centres; (2) that the convulsions, which may be due to various causes, are apt to be severe and accompanied by coma; (3) that the amaurosis may be associated with aphasia and paresis of hemiplegic distribution; (4) that the amaurosis is, for the most part, transient; (5) that in some instances there may be hemianopia.

(3) *Amaurosis following prolonged blepharospasm.*—That amaurosis may now and then follow long-continued intolerance of light and spasm of the eyelids in children suffering from phlyctenular ophthalmia is an observation that did not escape the attention of William Mackenzie. Writing in the year 1854 ('Practical Treatise on the Diseases of the Eye,' fourth edition, p. 486), that acute observer mentioned two cases of the kind, although apparently he was not aware that the blindness was of passing nature. The condition, which to be sure is very rare, may complicate blepharospasm in young children, irrespective of the cause of the latter. As soon as the eyes can be spontaneously opened, the child is found to be unable to recognise objects once quite familiar to him. The pupils

show no deviation from the normal, and there are no changes in the eye to account for the blindness. Sight is slowly regained in the course of several weeks, and during that period the child's general intelligence is duller than it was before the eyes got bad.

The following is a typical example of this rare condition :

(24) Emily O'D—, aged 3 years, was brought to the Evelina Hospital on October the 20th, 1899, suffering from bilateral interstitial keratitis of six weeks' duration. Her brother, aged 6 years, was under my care for a similar ailment. There had been three children born dead before the birth of my two patients. The keratitis in both cases was associated with an unusual amount of photophobia. Despite treatment, Emily's photophobia became so pronounced that on December the 29th, 1899, immersion was tried. On February the 23rd, 1900, photophobia still continuing, the lids were ordered to be opened with retractors for some minutes daily at the hospital. Three days later (February the 26th) the nurse who was carrying out this method of treatment discovered that Emily O'D— could see nothing, an observation afterwards amply confirmed by the child's mother. March the 23rd, 1900 —The child, who now opens her eyes pretty well, is stated to be unable to recognise objects once familiar to her. She seems dull-witted. On March the 27th, however, the child noticed a bright tin on the kitchen mantelpiece and exclaimed, "Hark at it shining!" During the next three days the child saw the fire shining, opened her eyes fairly, and ran into the street alone. April the 20th, 1900—Sight continues to improve. Child runs alone in the street and recognises common objects, as flowers, cups, saucers, jugs, glasses, and so forth. Both corneæ slightly cloudy, but no redness of the eyes or photophobia. Later in the history of the case the child developed bilateral dacryocystitis, associated with Staphylococcus albus and aureus in the discharge. The disease eventually yielded to treatment, specific and surgical.

To judge from the case about to be narrated, the amaurosis, the nature of which is still imperfectly understood, may be limited to one eye :

(25) A bright and talkative child, aged 2 years, developed on January the 20th, 1899, without known cause, cellulitis of the right orbit, accompanied by marked œdema of the lids, so that they could not be opened. The movements of the eyeball were abolished, and the globe itself was pushed forward and outward. The general symptoms were of a somewhat alarming character—child semi-conscious and in great pain, pulse uncountable, temperature 105° F. Three days later a deep incision was made through the upper lid into the orbit, and some pus escaped when dressing-forceps were introduced into the wound thus made. With a view to test the sight a week later (January the 30th), the lids of the affected eye were opened, the other eye being meanwhile kept covered up. As soon as this was done the child exclaimed, "Blind's down, daddy, all dark." The pupils were active, the media clear, and the ophthalmoscopic appearances normal. On another occasion when asked to see where the nurse stood, he said, "No; all dark." But when the good eye was uncovered, he at once exclaimed, "Why, there's Norah" (his nurse), who had been standing in front of him all the time. On February the 17th, the nurse noticed for the first time that the child could see her with his right eye, the other being closed. This observation was repeated the next day by asking the child what he saw in his mother's hat. "Flowers and feathers" was the correct answer to this question. It was also found that the little boy could name various objects in his night-nursery without mistake.

Leber's explanation of this singular condition, namely that it is due to cortical changes of a functional character, probably represents the truth. Broadly speaking, in such cases the child, who is always of tender age, may be said to have forgotten how to see, and before vision can be regained, he must re-educate his cortical centres.

Some Family Affections of the Eye.

Apart from conditions such as retinitis pigmentosa (with or without pigmentation of fundus) and Leber's optic neuritis or atrophy, which I shall not attempt to describe in this Lecture, there are three conditions of the eye seen in children which show a strong tendency, as it were, to run in families. They are—(1) amaurotic family idiocy; (2) progressive mental degeneration with changes in the fundi; and (3) degeneration of the cornea. The first is well known, but so far not a great amount of attention has been paid to the other two conditions.

(1) *Amaurotic family idiocy.*—In 1881 Mr. Waren Tay ('Ophth. Soc. Trans.,' vols. i and iv) described five instances of a curious condition in which characteristic changes in the fundus of the eye coincided with loss of muscular power in the neck and limbs. The five patients were derived from two families. Additional communications by Kingdon, Goldzieher, Magnus, Hirschberg, Wadsworth, and Hermann Knapp confirmed Mr. Tay's observations. In 1896 Dr. Sachs, of New York ('New York Medical Journal,' July, 1896), published an elaborate article on the disease, which he christened "amaurotic family idiocy," a name now generally adopted.

The disease, which is probably not an exceedingly rare one, affects, almost without exception, the offspring of Jewish parents. It is generally observed between the ages of 3 and 12 months in children who up to then have manifested no departures from the normal. The infant becomes apathetic, lies listlessly in his cot, and loses his sight. The ophthalmoscopic appearances are symmetrical and as striking as they are characteristic. They include (*a*) pallor of the optic discs, and (*b*) a milky-white area, having soft-looking edges and a red nucleus, which occupies the yellow spot region in each eye, and which recalls the appearances seen in embolism of the central artery of the retina. The eyes, now quite blind, roll aimlessly from side to side. Although the baby lies placidly in his bed, yet he is apt to jerk violently if a sudden noise be made. The rule is for the baby gradually to get worse, and, finally, to die,

paralysed and idiotic, within a year or eighteen months after the onset of the disease. At the same time, sudden death is not unknown. As pointed out before, the affection shows a pronounced tendency to attack several children belonging to the same family.

Sachs ('Journal of Mental and Nervous Disease,' January, 1903) concludes that the morbid process affects primarily, or at least to a great extent, the entire grey matter of the brain and spinal cord. Degeneration of the white fibres of the anterior and lateral pyramidal tracts is, in all likelihood, secondary. The relative involvement of the grey and white matter, however, may vary in different patients. Syphilis has probably nothing to do with the causation of the disease.

Prognosis is unconditionally bad.

As regards treatment, Kingdon and Russell suggest that children in whom the advent of the disease is to be apprehended should be fed artificially and not on the breast. The underlying idea is that something or other in the mother's milk may be responsible for the degeneration of the nervous system.

(2) *Progressive mental degeneration, with changes in the fundi.*—There exists a small but well-marked group of cases, occurring in young children, marked by eye changes, on the one hand, and by systemic changes, on the other. The former include slight pallor of the optic disc, smallness of the retinal vessels, and trivial pigmentary alterations situated chiefly in the yellow spot region of the fundus oculi. The general changes range from mere irritability and stupidity to actual idiocy.

The first reported observation with which I am acquainted was in 1897 by Dr. Rayner D. Batten, who showed at a meeting of the Ophthalmological Society two brothers, aged 14 and 21 years respectively, in whom the discs were pale, and there was an aggregation of fine stippled pigment at the macula. The condition of the fundus was similar and symmetrical in the two brothers. Mental defects, however, were absent. There was a definite history of syphilis in the parents, although the patients themselves presented no evidence of inherited disorder. In 1900 Dr. G. F. Still and Mr. Donald Gunn showed an analogous case in a child aged about 3 years. Marked muscular weakness, loss of knee-jerks, and nystagmus coincided with a grey area in the macular region of each eye. In 1902 Dr. F. E. Batten exhibited two cases in the same family, where the children were mentally defective and presented the associated fundus changes. In 1904 Professor Hirschberg published an isolated example of the affection in a young child. In the same year Mr. S. Mayou brought

under the notice of the Ophthalmological Society three members of a family, aged 10, 9, and 8 years, with both sight and mind affected. The parents of these children were themselves the offspring of the union between first cousins. Further, there was a suspicion of syphilis. Mr. Mayou summed up the chief points in his cases as follows: (1) A syphilitic history in the parents, but no other evidence of congenital syphilis in the children; (2) the simultaneous onset of the ocular and mental condition, generally between the ages of 5 and 14 years; (3) the loss of central vision; and (4) the typical appearance of the macula.

Additional cases of this type have been described by Dr. Robert Hutchison and by myself. In Hutchison's case, which occurred in a child, aged 4 years, an elder brother, aged 6 years, was completely imbecile.*

The symptom-complex of this type may be constructed somewhat as follows: A child, usually about the age of 5 or 6 years, who has previously been of normal development, perhaps after an attack of measles or other childish ailment, becomes irritable and subject to fits of temper, and grows dull mentally—"unteachable," as it were. He may develop spasticity of the legs, and exhibit more or less inco-ordination. The knee-jerks may be difficult to elicit or actually absent, and in some of the reported cases plantar stimulation has tended to give an extensor response (Babinski's sign). The sight is noticed to be defective, and the child looks at objects held in front of him sideways, thus manifesting defect in his powers of central vision. The general examination of such a patient shows that he is very backward and extremely dull and often extremely timid, so that he is apt to burst into tears almost without provocation. The reaction of the pupils, which are apt to be moderately dilated, is defective, and nystagmus or defects in the movements of the eyes, as in a case of my own to be related immediately, may be present. When the ophthalmoscope is used, slight pallor of the optic disc is found, the retinal vessels may be somewhat small—"on the small side" in hospital slang—and the yellow spot region shows slight disturbances, characterised by irregular aggregation of retinal pigment—a slight form of choroido-retinitis, in fact. The pigmentary disturbances, however, need not be limited to the macula, but may affect the central part of the fundus, and perhaps an even wider area, but they are never, so far as I know, pronounced at the periphery, as is the case in the ordinary choroiditis of hereditary syphilis.

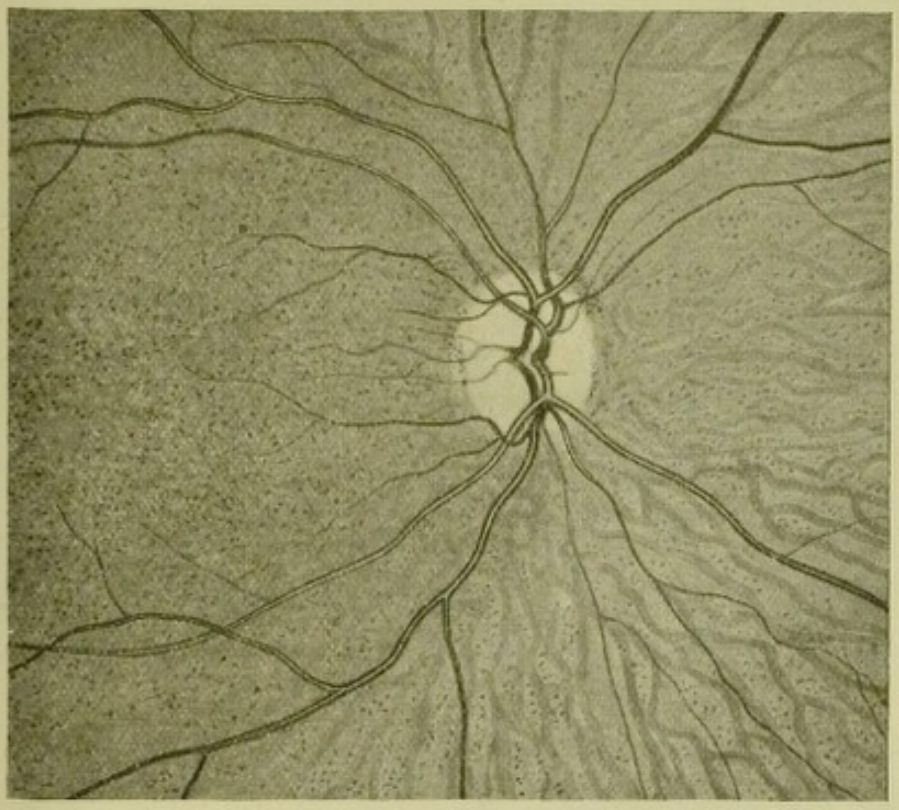
* A case of Dr. Kauffmann's ('Ophthalmoscope,' September, 1905) may belong to the same category.

There is no tendency in this affection, as in amaurotic family idiocy, to attack Jewish children almost exclusively.

The disease slowly becomes worse, and culminates in total central blindness and in complete imbecility. But while this appears to be the usual outcome, yet in rare instances, as in a case of my own, to be next described, improvement may take place under treatment:

(26) Albert B—, aged 7½ years, was first seen at the North-Eastern Hospital for Children on May the 19th, 1904. He was the only child. His parents were not related by blood-ties. They had been married for nine years, and there had been no mis-

Fig. 7.



carriages or stillbirths. According to the history given, Albert had been a bright lad until about twelve months before he fell under my notice, but then his sight began to fail, he became very nervous and intensely stupid, and his memory failed, so much that he had to be taken away from school. He had suffered from bronchitis, German measles, and influenza at 4½ years, from measles at 5 years, from chickenpox shortly afterwards, and, finally, from mumps at 6½ years. *Upon examination.*—The child is noticed to stare about him in a fixed and sightless sort of way. He can scarcely raise his eyes above the horizontal plane, and lateral movements, also, are limited. Some night-blindness exists. Slight pigmentary changes are present in the macular region of each fundus. The knee-jerks and the superficial reflexes are present, but the plantar reflex is of the extensor type. No personal dystrophic stigmata of syphilis. *Treatment and progress:* The patient was placed upon potassium iodide, two grains three times a day. In August, 1904, some little improvement having taken place, vision was ascertained to be $\frac{1}{60}$ and No. 19 Jaeger. My colleague, Dr.

James Taylor, who was good enough to examine the lad, reported that there was "no obvious sign of structural nervous disease." In October, 1904, a note was made to the effect that the pupils were equal and active, and that the child's general condition had improved. He could read No. 14 Jaeger with either eye, and recognise objects such as a penny, a box of matches, a brush, and so forth, at a distance of two feet. He always fixed to the right of an object held before him. The downward movements of his eyes are good, but all the other movements are defective. Hippus. The discs, especially as regards the outer half, a little pallid. Other fundus changes as noted. In April, 1905, the lad was thought to be getting brighter. He then read No. 20 Jaeger. Upward movement of the eyes was absent. I saw Albert B— the other day (July the 6th, 1905) and made the following note: "The optic discs, especially the left one, pallid. The fine pigmentary changes are not now limited to the macular region, but are also present in the circumpapillary region of the fundus, but not in the periphery. Upward movement of the eyeballs can scarcely be said to exist. General condition better."

My excuse for having dwelt at some length upon this form of mental and ocular degeneration is because the type has hardly yet been crystallised into concrete and tangible form, although individual cases must be familiar to every ophthalmologist and to most physicians.

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(3) *Degeneration of the cornea.*—It has been known for some little time that, in rare instances, a peculiar degenerative affection of the cornea may be transmitted in families. This condition may be fairly included under what Sir William Gowers has recently termed "abiotrophy"—that is to say, a peculiar tendency in certain structures, particularly in those of the nervous system, to premature decay and to early death.

Under the name "lattice-like" keratitis, Professor Haab (1) described, in 1899, this curious condition in a boy, aged 16 years, whose maternal uncle and aunt were also affected. The corneal opacities looked at first glance as if they were the residue of an interstitial inflammation, but closer examination resolved them into a lattice-like system of lines intermingled with fine rounded dots. Symptoms of inflammation were conspicuous by their absence. Similar cases have

been published, among others, by Dimmer (2), Hauenschild (3), and Freund (4). The last-named surgeon was able to refer to fifteen cases belonging to two families, of which one furnished seven and the other eight patients. In England this form of disease has been reported by R. Marcus Gunn (5) and W. T. Holmes Spicer (6).

Under the name of "nodular" opacities of the cornea another form of disease, which occurs under conditions similar to that just described, has been mentioned by several writers, as Groenouw (7), Manz (8), Eversbuch (9), Neznamow (10), Fuchs (11), R. W. Payne (12), Oscar Dodd (13), E. T. Collins (14), Koerber (15), and Deutschmann (16). The condition, which is associated with no signs of inflammation, takes the form of discrete grey dots, occupying at first almost exclusively the central region of the cornea.

Fig. 8.

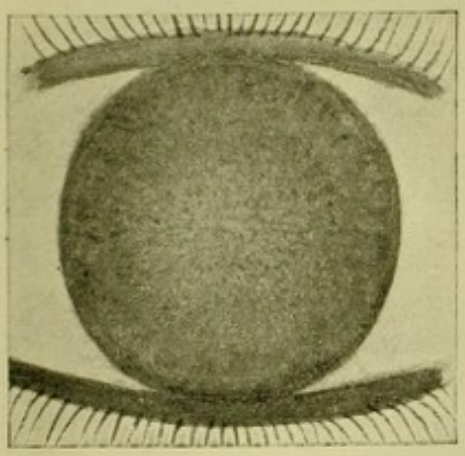
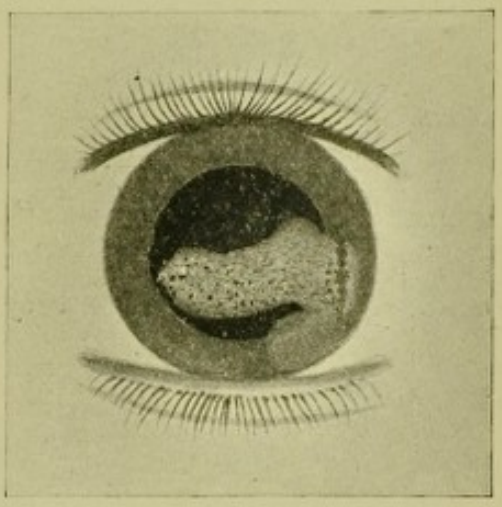


Fig. 9.



These two corneal degenerations, "lattice-like" and "nodular," therefore agree in several particulars. For example, they generally begin at about the age of puberty, they are slowly progressive, they attack at first the central parts of the cornea, and, lastly, they show a marked tendency to run in families. The conclusion is that, at root, they are merely varieties of one and the same process of abiotrophy. This view is strengthened by a series of cases published recently by Mr. R. W. Doyne and myself (17), in which three generations of a particular family manifested seven instances of a slowly progressive and bilateral degeneration of the cornea, leading to serious impairment of sight after lasting for several years. Neither syphilis nor consanguinity existed. The thyroid gland showed no changes. Some of the patients presented the so-called "nodular" and others the "lattice-like" opacities, while in still a

third group the corneal changes did not resemble either of the two conditions named (see figures 8–12). At the same time, it would be opposed to sound reason to suppose that the malady in its essential features differed in the various members of the family described by Mr. Doyne and myself.

Fig. 10.

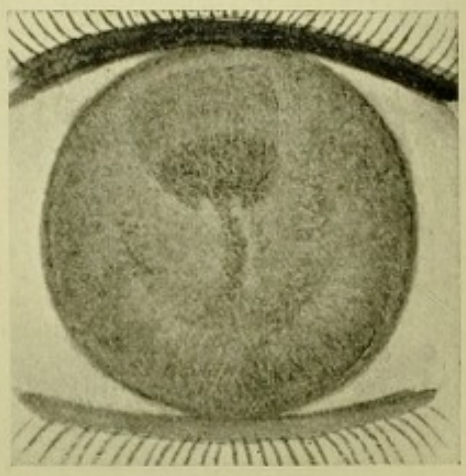
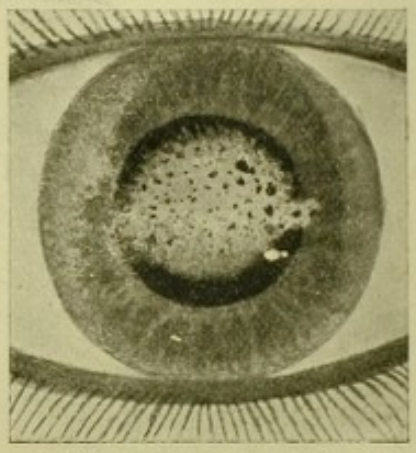
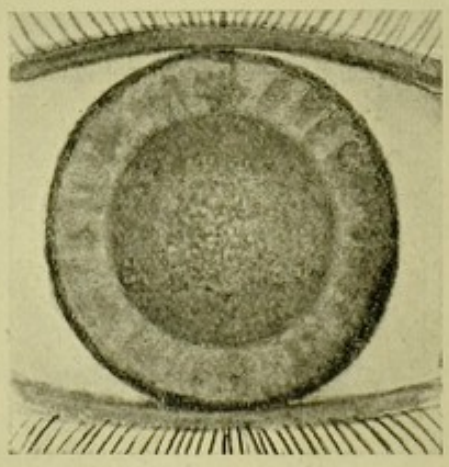


Fig. 11.



We may reasonably conclude that there exists a "family degeneration of the cornea" of a special kind that may assume clinically either the "nodular" or the "lattice-like," or some intermediate and indeterminate type.

Fig 12.



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Congenital Word Blindness.

Thanks to the labours of James Kerr, W. Pringle Morgan, Bastian, Hinshelwood, Nettleship, Lechner, Wernicke, and Bruner, surgeons have during recent years become familiarised with a singular congenital defect in which otherwise intelligent children experience more or less difficulty in learning to read. The number of reported cases is as yet small, but there are now substantial reasons for believing that congenital word blindness is far from uncommon, especially among the children of the lower classes.* Eighty-five per cent. of the cases have been reported by ophthalmic surgeons, a fact that goes to show that among the better classes such patients, at all events in the first instance, seek special rather than general medical advice. The disability differs in intensity, and ranges from a mere difficulty in learning how to read to an almost complete inability to do so. On the other hand, arithmetical numerals seldom offer any difficulty. A majority of the published cases have been in males, a point in which word blindness reminds one of colour blindness. Some of the sufferers have been of a markedly neurotic temperament, as in cases published by myself. I have recently met with two cases (still unpublished) in which a similar defect had existed in the mother or in other members of the family.

The condition is probably due to a congenital defect in the visual memory centre for words and letters, and it will be a matter of great interest to inquire into its relationship with allied states, such as congenital word deafness, amusia, ecolalia, "word cropping," and idioglossia. The auditory memory has been highly developed in

* Dr. C. J. Thomas ('Ophthalmoscope,' August, 1905) calculates that 1 in 2,000 of all London Elementary School children are congenitally word blind, and he states that there are now nearly one hundred instances of the condition noted in the case-books at the special schools for the mentally defective. His original communication should be read by all interested in the subject.

most of the patients. One of Wernicke's patients could speak two languages correctly; a patient of Hinshelwood's concealed his difficulty in reading for a time by learning the contents of his primer off by heart; Lechner's patient is stated to have known the contents of his school books by rote; and one of my own patients could memorise well and quickly anything told or read to him. Indeed, not the least interesting point about the condition is the way in which children compensate for and often manage to conceal their defect. They get help from the auditory route and especially from the so-called kinæsthetic memory—*i. e.*, the repeating of a sentence with the lips.

The improvement to be obtained in these children depends obviously upon the original severity of the case and the educational means adopted. With individual tuition even severe instances of alexia are capable of considerable improvement. It is clear that the sooner the defect is recognised the earlier can such training be commenced. Education in these children should be individual and largely verbal.

Diagnosis offers few difficulties if once the fact be grasped that there is such a thing as a congenital inability to read. Confusion, however, might arise when word blindness co-existed with a considerable error in refraction. Even then the history should safeguard us against error, while the fact that the child experienced exactly the same difficulty in reading large as he did small type would be strongly suggestive of the existence of this curious and interesting congenital defect. It may be added that more than one of the case histories comment upon the fact that the patient never read for pleasure.

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Injuries to the Eye During Labour.

It is a matter of familiar knowledge that during the progress of labour, natural or instrumental, the eyes of the child are liable to be

injured. Such injuries, in addition to being of the most varied kinds, may be brought about in several different ways, of which the most important are the application of forceps, clumsy digital examinations, and prolonged labour in women with deformed or unusually narrow pelves. Some of the injuries are of little practical importance, as the common and harmless effusion of blood beneath the conjunctiva; whereas others, as dislocation of the eyeball, are of the most serious description. Leaving on one side such injuries as ecchymosis or wounds of the eyelids and fractures of the orbit, we have to consider four particular injuries, viz.—(1) corneal lesions; (2) retinal hæmorrhages; (3) dislocation of the eyeball; and (4) obstetrical paralysis.

(1) *Corneal lesions.*—A valuable article by Drs. Thomson and Buchanan (1) upon certain injuries to the cornea now and then sus-

Fig. 13.

tained by the infant during birth has focussed the attention both of ophthalmic surgeons and of obstetricians upon these lesions. Cases of the kind have been reported by Noyes (2), Dujardin (3), de Wecker (4), Truc (5), Sidler-Huguenin (6), Cargill (7), Fejer (8), Wolff (14), and myself (9). Drs. Thomson and Buchanan, however, have seen a considerable number of these cases in Glasgow, where rickets and consequently deformed pelves appear to be singularly common. How rare the condition is in some other places is shown by our experience at Queen Charlotte's Hospital, London, where on the average 1200 births take place annually, and forceps are applied in fully 10 per cent. of the births. At that hospital one case of injury to the cornea only has been recognised.

According to Thomson and Buchanan, there are two forms of corneal opacity—(1) a diffuse temporary opacity, and (2) a permanent linear opacity, more often multiple than single. The first

form (much the commoner) is due to œdema. The second form is the outcome of rupture of the posterior elastic lamina of the cornea and the subsequent formation of fibrous tissue. The common appearance will be seen from the figure, sketched from a case under my care.

Now, a majority of these injuries have followed instrumental labour, and are presumably due to direct injury by the forceps. This explanation (which has, at least, the merit of simplicity) does not account for every case, since Sidler-Huguenin and Thomson and Buchanan have published instances where it was evident that no such injury was likely to have taken place. For example, bilateral œdema of the cornea was produced in one baby (stillborn) by blocking of a normal pelvis by the simultaneous presentation of the face, arm, and cord, and in this case instruments were not employed. Besides, it must be remembered that forceps are applied only in difficult cases, and that the injury may be due, not to the instrument at all, but to the cause or the causes of the difficult labour. From a medico-legal standpoint it is clear that the possibility of non-instrumental injury is important.

As regards the permanent results of these injuries there can be no doubt, as in a case of my own, that the scar-tissue produces considerable irregular astigmatism and consequent interference with sight. Another point is that confusion may arise between these injuries, on the one hand, and so-called "congenital opacity" of the cornea, on the other.

(2) *Retinal hæmorrhages.*—The frequency of retinal hæmorrhages after normal labour has been estimated at from 10 per cent. to 32 per cent. (Schleich). Coburn (10) examined the eyes of thirty-seven infants, either stillborn or who had survived not longer than 22 days. Hæmorrhages were found in seventeen instances—that is, in 45 per cent. Most of the ecchymoses lay in the equatorial part of the eye, while the macular region presented few such changes. In other words, Coburn's histological investigations go to show that hæmorrhage may often be present, even although to ophthalmoscopic examination the fundus looks healthy. It is suggested that the extravasations result from disturbance of the retinal circulation, due to compression of the central blood-vessels of the optic nerve by cerebro-spinal fluid under high pressure. They are therefore to be regarded as an evidence of intra-cranial pressure leading to blood stasis. Wehrli (13) has recently suggested that in a majority of cases the exciting cause of glioma of the retina is be found in retinal hæmorrhage sustained at birth.

(3) *Dislocation of the eyeball.*—Cases of dislocation of the eyeball, partial or complete, have been reported in this country by Beaumont, Snell, and by Thomson and Buchanan. In most of the cases there has co-existed a depression of the cranial bones above the orbit on the affected side, so that Thomson and Buchanan are undoubtedly correct in designating the injury as "extrusion" of the eyeball rather than "dislocation." There is another possible explanation, as in a celebrated case quoted by de Wecker (4), where an accoucheur, mistaking a face for a buttock presentation, managed to gouge out the baby's eye with his finger. Short of actual extrusion of the eyeball, cases are known in which there has been proptosis, or in which the optic nerve has been torn through.

(4) *Obstetrical paralysis.*—The form of obstetrical paralysis most familiar to practitioners is perhaps that of the facial nerve, due to compression or bruising by the forceps or by the pelvis of the nervous trunk where it emerges from the stylo-mastoid foramen of the temporal bone. It does not appear to be so generally known, however, that several other cranial nerves may suffer, individually or collectively, as, for example, the third, fourth, and sixth. Even the superior cervical ganglion of the sympathetic may be involved, to judge from a case reported by Reese, who observed in a baby delivered by forceps cranial deformity, narrowing of the palpebral fissure, contraction of the pupil, and enophthalmos.

The exact mechanism whereby these various paralyses are produced is not always very evident, although it is tolerably clear, as hinted, in the case of facial paralysis. As regards the third, fourth, and sixth nerves, it may be due either to fracture of the bones of the orbit or to intra-cranial or intra-orbital extravasation of blood. The fact that the paralysis not infrequently is of a more or less transient character favours the last-named view. In a case described by Küstner, where the third, fourth, sixth, and seventh pairs were affected, the paralysis was found after death to be due to hæmorrhages at the base of the brain, as well as to lesions of the orbital walls.

The subject of obstetrical paralysis gains added importance from the fact that such injuries may be confused with so-called "congenital paralysis" of the ocular muscles, due either to the maldevelopment of centres or of muscles, or more probably, as I now think, to intra-uterine polio-encephalitis.

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